Complex Congenital Heart Disease in Adults
Linda B. Haramati, MD

Learning objectives
- List the main underlying anomalies in tetralogy of Fallot and transposition of the great arteries
- Learn the major surgical treatments for these anomalies
- Describe the roles of CT and MR in guiding management for adults with tetralogy of Fallot and transposition of the great arteries

Complex Congenital Heart Disease in Adults
- Neonatal surgery 1970s markedly improved survival of children born with CHD
- ~85% surviving 1 million adults with CHD
- Adults with complex CHD should be treated in specialized centers
- Suffer from complications of longstanding CHD and its treatment

Complex Congenital Heart Disease in Adults
- Tetralogy of Fallot (TOF)
- Transposition of the great arteries (TGA)

TOF
- 6.8% of CHD, most common cyanotic CHD
- Described in 1888 by Fallot
- Malalignment of the conal septum in relation to the ventricular endocardial cushion
- Malaligned VSD, overriding aorta & RVOT obstruction (RVH)
- Disease severity depends on degree of RVOT obstruction
- Pulmonary atresia in 20%

Disclosures
Family member
- Board Member & Shareholder
  - Kryon Systems, LTD
  - Orthospace, LTD
  - Bioprotect, LTD
- Executive Advisory Board
  - GE Healthcare IT
• 1950s: initial surgical repair
• VSD closure & relief of RVOT obstruction
  > Tranannular patch, RV-PA conduit, infundibulectomy
• Unique problems related to longstanding postoperative physiology & delayed consequences of disease
• Prolonged post-op pulmonary insufficiency
  > Nearly universal & most common imaging indication

---

**Imaging Modalities**

- **Echocardiography** – mainstay
- **MR** - specific indications anatomic & quantitative
- **CT** - when MR contraindicated
  > coronary artery anatomy
  > pulmonary parenchymal assessment

---

**MR protocol**

<table>
<thead>
<tr>
<th>MR protocol</th>
<th>Information gleaned</th>
</tr>
</thead>
<tbody>
<tr>
<td>ECG-gated black double IR</td>
<td>Anatomic overview</td>
</tr>
<tr>
<td>SSFP cine - short axis, long axis, RVOT</td>
<td>RV, LV volumes, EF, wall motion, mass, valves</td>
</tr>
<tr>
<td>Phase contrast MPA (RPA, LPA), aorta</td>
<td>Flow quantification, velocity</td>
</tr>
<tr>
<td>C+ MRA coronal</td>
<td>Central &amp; peripheral PA anatomy</td>
</tr>
<tr>
<td>Delayed enhancement</td>
<td>Fibrosis</td>
</tr>
</tbody>
</table>

---

**Case 1: s/p TOF repair in infancy**

Shortness of breath on exertion

- **Echo** - free PI with RV dilation, tricuspid regurg
- **MR** - RV volumes, regurgitant fraction
What does this mean?

- PI initially well-tolerated post repair
- Over decades, morbidity & mortality from chronic PI→RV dilation, TR in 10%, ultimately... biventricular dysfunction, failure & arrhythmia
- PVR standard practice in symptomatic pts, improved PI, volumes & sx but not RVEF
- ? impact on mortality
- Optimal timing of PVR
  - under investigation

Case 1 - cont’d

- RX-PVR and tricuspid valve annuloplasty
- MR performed 1 & 2 years postoperatively

Quantitative Analysis

<table>
<thead>
<tr>
<th></th>
<th>Indexed RVEDV</th>
<th>Indexed RVESV</th>
<th>RVEF (%)</th>
<th>Regurg. Fraction (%) at PV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Index MR</td>
<td>156 ml/m2</td>
<td>106 ml/m2</td>
<td>32</td>
<td>53</td>
</tr>
</tbody>
</table>

Quantitative Analysis

<table>
<thead>
<tr>
<th></th>
<th>Indexed RVEDV</th>
<th>Indexed RVESV</th>
<th>RVEF (%)</th>
<th>Regurg. Fraction (%) at PV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Index</td>
<td>156</td>
<td>106</td>
<td>32</td>
<td>53</td>
</tr>
<tr>
<td>* surg</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Postop</td>
<td>112</td>
<td>84</td>
<td>25</td>
<td>21</td>
</tr>
<tr>
<td>1 year later</td>
<td>114</td>
<td>80</td>
<td>30</td>
<td>21</td>
</tr>
</tbody>
</table>
Increased mortality
MRI: RV mass/volume ratio >0.3 g/ml (RVH) & decreased LVEF (or RVEF)

CT in TOF
- Ventricular volumes, wall motion and ejection fractions - retrospective ECG-gating
- Inferentially calculate regurgitant volume if isolated PI (no TI or VSD) based on equal right and left heart cardiac outputs
  - RV SV-LV SV = pulmonary regurgitant volume
- Imaging modality of choice for coronary anomalies, pulmonary parenchyma

Case 2:
20-year-old man S/P TOF repair
RV-PA conduit
- Single coronary artery
- B-T shunt takedown at definitive repair
- S/p pacemaker
- CT for ventricular volumes, coronary and PA anatomy
TOF with pulmonary atresia Spectrum

- Large confluent central PAs discontinuous but near RVOT
- Absent central PAs with lungs supplied solely by systemic collaterals (pseudotruncus)
- Numerous intermediate forms
- Primary repair for milder forms
- Absent central PA’s difficult to repair (unifocalization)
- MR and CT- central PAs, systemic collaterals

TOF- lungs

- Lung function often abnormal after repair
- Diminished perfusion
- Central & peripheral PA stenoses
- PA stenosis exacerbates PI → increase RV ESP & decreased exercise tolerance
- TOF with PA, lung segments supplied exclusively by nonstenotic collaterals develop pulmonary HTN

CT-local vasculature

- Perfusion: adequate, under or over
- Mosaic perfusion
- Systemic collaterals directly visible or serrated pleural parenchymal bands

20 F TOF s/p repair absent LPA

Anatomy of TGA (van Praaghe)

- Normal: (S, D, S) – atrial situs solitus, ventricular D-looping and normally related great arteries (2 circuits in series)
  Atrioventricular concordance, ventriculoarterial concordance
  Aorta is posterior and to the right of the PA (crossed orientation)
- Situs solitus and complete transposition of the great arteries (D-TGA): (S, D, D) (2 circuits in parallel)
  Atrioventricular concordance, ventriculoarterial discordance
  Aorta is anterior and to the right of the PA (parallel orientation)
- Situs solitus and congenitally-corrected transposition of the great arteries (L-TGA): (S, L, L) (2 circuits in series)
  Atrioventricular discordance, ventriculoarterial discordance
  Aorta anterior and to the left of the PA (parallel orientation)
Complete (D-TGA): 2 circuits in parallel

- Neonatal cyanosis
  - With PDA closure
- Obligate shunt for life
  - VSD, ASD
- Atrial septostomy
  - Early complete repair
  - Atrial switch
    - Senning (1957)
    - Mustard (1964)
  - Rastelli
  - Arterial switch
    - Jatene (1975)

Atrial switch for D-TGA

MR (CT) for D-TGA / Atrial switch

- Baffles: Systemic (SVC, IVC) & Pulmonary venous pathways
  - Obstruction
  - Leak: Qp:Qs
- Ventricular volumes & function
  - Systemic RV
- Valves

Failure of the systemic RV

RV-EDV 132ml
RV-ESV 108ml
RVEF=18%

Arterial switch for TGA
MR (CT) for D-TGA / Arterial switch

- Pulmonary artery anatomy
- Coronary artery anatomy
- Ventricular function
- Ventriculoarterial valve function
- Fibrosis

Conclusion: Complex CHD in Adults

- Early repair of complex CHD → improved survival and growing adult population
- TOF repair → pulmonary insufficiency
  > well-tolerated early, but associated with morbidity & mortality
  > MR - ventricular volumes & function, PI, PA anatomy and flow, fibrosis
  > CT - MR contraindicated, coronary anatomy, lungs

Conclusion: Complex CHD in Adults

- D-TGA – early presentation and treatment
  > Atrial switch - systemic and pulmonary venous limbs of baffle for obstruction, baffle leak, systemic RV function
  > Arterial switch - outflow tract & PA stenosis, coronary stenosis, ventricular and valve function

Thank you